

Persistent left fifth aortic arch in man

Report of two cases

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Two cases of persistent left fifth aortic arch forming a congenital double lumen aortic arch have been confirmed at necropsy. In the first case, the diagnosis was suspected during life after aortography. An unusual vessel ran inferior and parallel to the aortic arch from the innominate artery to the left subclavian artery. It lay superior to the pulmonary artery.

Both cases were associated with other cardiovascular anomalies. The first case had a persistent ductus arteriosus, and a small membranous ventricular septal defect; the second, coarctation of the aorta, persistent ductus arteriosus, bicuspid aortic valve, and a single right coronary artery.

There is disagreement about the existence of the fifth aortic arch in man. In the cat embryo, Huntington (1919) depicts a fifth arch forming a vascular channel beneath the fourth arch. Brown (1913) in the cat embryo, and Buell (1922) in the chick embryo have described the existence of the fifth arch. Congdon (1922) reported that endothelial sprouts arising from the aortic sac ventrally and from the descending thoracic aorta correspond to the upper and lower ends of the fifth arches in man. Sissman (1968) and Hamilton, Boyd, and Mossman (1962) allege that the status of the fifth aortic arches is uncertain. Balinsky (1965), Arey (1965), Langman (1969), and Duckworth (1967) either deny the existence of the fifth aortic arch in mammals, or believe that it is only present transiently if it does exist.

Van Praagh and Van Praagh (1969) described the first case of duplication of the aortic arch and suggested that the smaller caudal arch or 'subway' represented the fifth aortic arch. In this presentation, we describe, to the best of our knowledge, the first case of a persistent fifth aortic arch diagnosed during life and a third example of this rare malformation in which there were two complete aortic arches of almost equal size arising anterior to the trachea.

Case reports

Case 1

A female infant, 4½ months old at the time of death, was born prematurely at 32 weeks' gestation weighing 1705 g

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after an assisted breech delivery. The Apgar score was 1. Intensive resuscitation was necessary. Initially, clinical findings and chest x-ray were compatible with the diagnosis of respiratory distress syndrome. Respiratory difficulty persisted, and from the end of the first week she required assisted ventilation by intermittent positive pressure. She also had convulsions due to hypoglycaemia and hypocalcaemia.

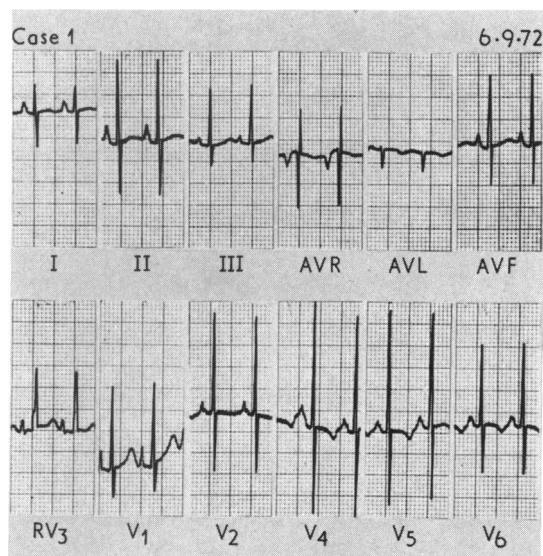


FIG. 1 Case 1. Electrocardiogram with evidence of right atrial hypertrophy and combined ventricular hypertrophy.

In the second week of life a systolic murmur was noted along the left sternal border and she developed congestive cardiac failure. A chest x-ray at this time showed cardiomegaly and pulmonary plethora. An electrocardiogram showed right atrial hypertrophy and combined ventricular hypertrophy (Fig. 1). Clinically a diagnosis was made of persistent ductus arteriosus and/or ventricular septal defect with pulmonary hypertension. She was treated with digoxin and diuretics.

The infant showed an inability to swallow and abnormal neurological responses. Tube feeding was required throughout her life because of aspiration whenever an attempt was made to bottle feed. The dysphagia was attributed to neuromuscular disorder. A barium swallow failed to show any compression on the oesophagus from a vascular ring. An electroencephalogram showed non-specific and generalized abnormal changes.

At the age of 3 months, a cardiac catheterization was

carried out because of the persisting heart failure and cardiomegaly. The presence of a moderately large persistent ductus arteriosus and a small ventricular septal

TABLE Cardiac catheterization data

Site	O ₂ Saturation (%) (breathing 28% O ₂)	Pressure (mmHg)	
		Phasic	Mean
Superior vena cava	61	a = 15	
Right atrium	52.5	v = 11	6
Inferior vena cava	66		
Right ventricle	73	75/8	
Pulmonary artery	82	75/34	50
		a = 10	
Left atrium	92	v = 12	8
Pulmonary vein (left)	92		
Left ventricle	91	94/9	
Descending aorta	93	95/50	62

Approximate pulmonary to systemic blood flow ratio = 2.9:1

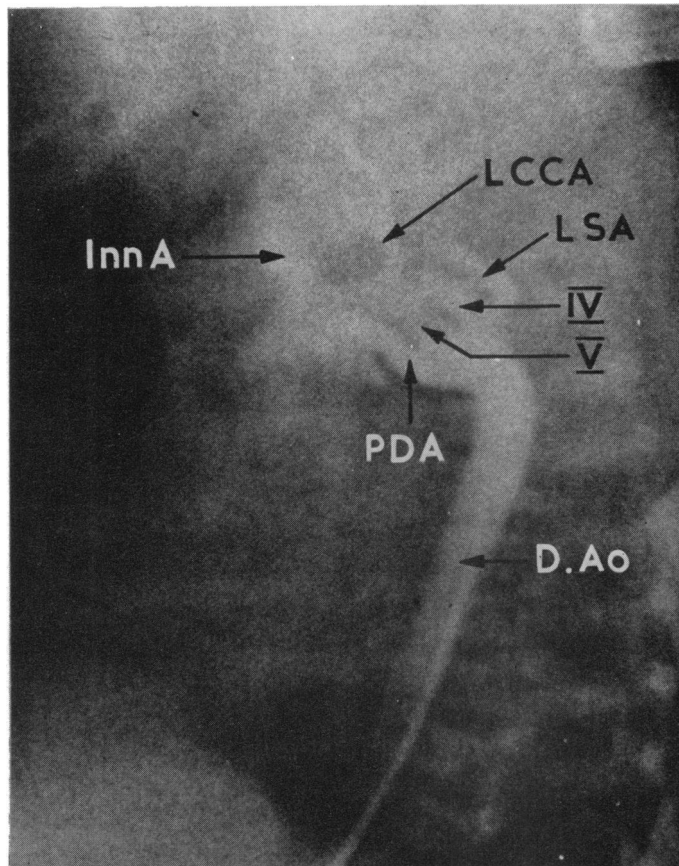


FIG. 2 Case 1. Aortogram. Three parallel horizontal channels are seen in this lateral view. From the upper fourth aortic arch (IV) arise from front to back, the innominate artery (Inn A), the left common carotid artery (LCCA), and the left subclavian artery (LSA). The middle channel is the persistent left fifth aortic arch (V), and the lower channel is the persistent ductus arteriosus (PDA). The catheter lies in the descending aorta (D. Ao).

defect was confirmed (Table). There was a 12 per cent rise in oxygen saturation from the superior vena cava to the right ventricle and a 9 per cent rise from the right ventricle to 82 per cent in the pulmonary artery. The pulmonary venous and systemic saturations were 92 and 93 per cent, respectively. The pulmonary to systemic flow ratio was approximately 2.9:1.

There was pulmonary hypertension with the main pulmonary artery pressure at 75/34 mmHg (mean 50 mmHg). The aortic pressure was 95/50 mmHg (mean 62 mmHg).

A left ventricular angiogram revealed a small aneurysm of the membranous portion of the interventricular septum with a small defect at the apex of the aneurysm. An injection of contrast medium into the

persistent ductus arteriosus opacified the pulmonary artery and descending aorta. There was also retrograde filling of the aortic arch, and an unusual vessel between the aortic arch and the persistent ductus arteriosus was noted. An aortogram revealed a normal left aortic arch with normal brachiocephalic arteries and a persistent ductus arteriosus in the normal position. Again, a separate channel running parallel to the undersurface of the arch was seen arising from the aorta just beyond the origin of the innominate artery to rejoin it at the level of the left subclavian artery (Fig. 2) (Moes and Izukawa, 1973, unpublished observations).

Shortly thereafter, the persistent ductus arteriosus, which measured 4 mm in diameter, was surgically ligated. Dissection along the aortic arch revealed broadening and

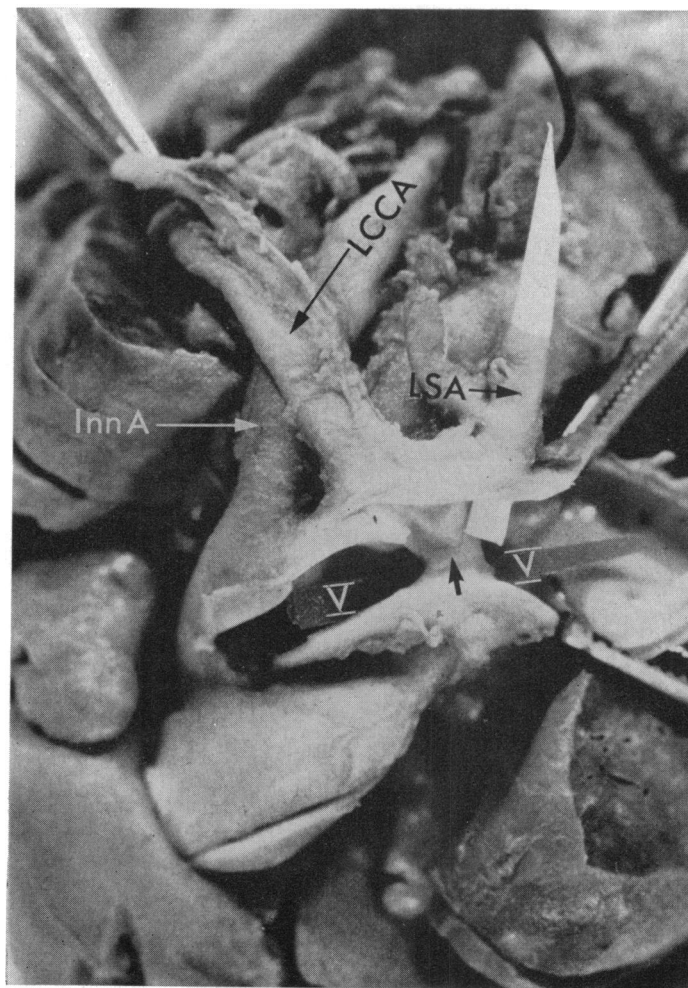


FIG. 3 Case 1. Specimen to show the two aortic arches; the upper channel has been opened to expose the bar of tissue (\uparrow) separating the left fifth aortic arch situated posteroinferiorly (V). The innominate artery (Inn A), left common carotid artery (LCCA), and left subclavian artery (LSA) arise from the left fourth aortic arch.

flattening from the level of the innominate artery to the left subclavian artery.

The infant survived operation, but had recurrent aspiration pneumonias and died at 4½ months of age.

Necropsy There was an absence of thymic tissue grossly and on microscopy.

The ascending aorta was 20 mm in internal circumference as compared to the larger main pulmonary trunk of 30 mm in circumference. The great arteries were normally related. The brachiocephalic arteries arose from the dorsal aspect of the left aortic arch in the usual order of innominate artery, left common carotid artery, and left subclavian artery. The aortic arch at the innominate artery level measured 15 mm in internal circumference and the descending aorta also measured 15 mm in circumference. Internally, the aortic arch channel from beyond the origin of the innominate artery to the left subclavian artery was divided into an upper and dorsal channel (measuring 4.5 mm in internal diameter approximately) and a lower and ventral channel measuring 3.5 mm in internal diameter. The channels, which were symmetrical, were separated by a horizontal bar of tissue from anterior to posterior aortic wall along the apex of the arch (Fig. 3). The diameter of the aorta was increased at the site of the aortic end of the previously ligated ductus arteriosus.

The aortic valve and coronary arteries were normal. The interventricular septum was intact and there was no trace of the defect in the membranous septum, which had closed spontaneously. The foramen ovale was patent as a 4 mm slit anteriorly.

There were scattered areas of bronchopneumonia, atelectasis, and haemorrhage in the lungs. The brain was oedematous with softening and cystic formation anteriorly along the lateral ventricles.

Case 2

A male newborn, weighing 2900 g, was admitted to the hospital 7 hours after delivery. He was the fifth child of a 42-year-old woman who had had a severe attack of 'influenza' during the eighth week of pregnancy.

In the third trimester she developed hydramnios and the membranes had ruptured during the 36th week of pregnancy. Labour was induced 7 days later because of foetal distress. One minute after delivery the Apgar score was 3. The baby required resuscitation and was deeply cyanosed until placed in 70 per cent oxygen.

Physical examination revealed a slightly cyanosed infant. The face was peculiar, with small eyes, low-set ears, and a right-sided facial palsy. The neck was short and the left hand malformed. The heart rate was 160/minute and the respiratory rate 46/minute with sub-costal retraction. The liver edge was 2 cm below the costal margin and the tip of the spleen was palpable. The femoral pulses were very weak. The praecordium was hyperdynamic. The first heart sound was normal; the second was loud in both aortic and pulmonary areas and splitting was not discernible. A grade 3/4 ejection systolic murmur was audible over the lower praecordium and in the left axilla.

The chest x-ray showed a large heart with a cardio-thoracic ratio of 65 per cent. There was slight accentuation of the vessel markings. The electrocardiogram (Fig. 4) showed sinus rhythm, and hypertrophy of both atria and right ventricle; the mean frontal QRS axis was $+140^\circ$ and the T wave axis was -15° .

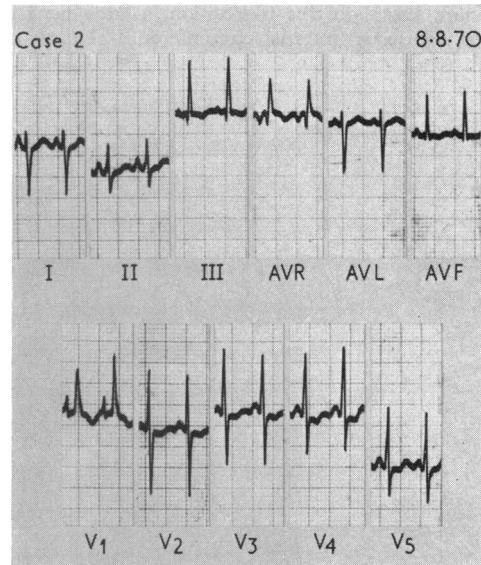


FIG. 4 Case 2. Electrocardiogram reveals right axis deviation of the frontal QRS axis (140 degrees), right atrial hypertrophy, and right ventricular hypertrophy.

The clinical diagnosis was heart failure due to coarctation of the aorta, probably associated with a more complex cardiac malformation. Pulmonary infection was considered a probability and cerebral damage a possibility. The infant was treated in an incubator with 40 per cent oxygen, intramuscular digoxin, frusemide, and vitamin K, and intravenous antibiotics and sodium bicarbonate. Before further investigations could be performed the baby died.

Necropsy Examination of the heart revealed slight right ventricular hypertrophy. No septal defects were present, but the foramen ovale was probe patent. The aortic valve was bicuspid with no discernible raphe to represent a third commissure. A single coronary artery arose from the right anterior sinus of Valsalva. The origin of the absent left coronary artery was represented only by a shallow dimple on the floor of the remaining sinus of Valsalva. No coronary artery arose from the pulmonary artery.

The ascending aorta measured 13 mm in internal circumference and was normally related to, but much smaller than, the main pulmonary artery (internal circumference 25 mm). At the origin of the innominate

artery, the aortic arch divided into two separate channels of equal size (internal circumference of 6 mm in each case: Fig. 5). The left common carotid artery and left subclavian artery arose, respectively, from the middle and terminal portions of the upper aortic arch. Immediately distal to the point where the two arches reunited there was a deep indentation in the left side of the aorta, at which point the internal circumference was reduced to 4 mm. Distal to this coarctation, a large persistent ductus arteriosus (internal circumference 16 mm) con-

nected the pulmonary artery and descending aorta (Fig. 5).

Histological examination confirmed the two arches to be entirely separate, each having its own adventitial sheath.

Discussion

In the mammal, there is clear embryological evidence of the existence of a left fifth aortic arch. In the cat embryo, Huntington (1919) and Brown

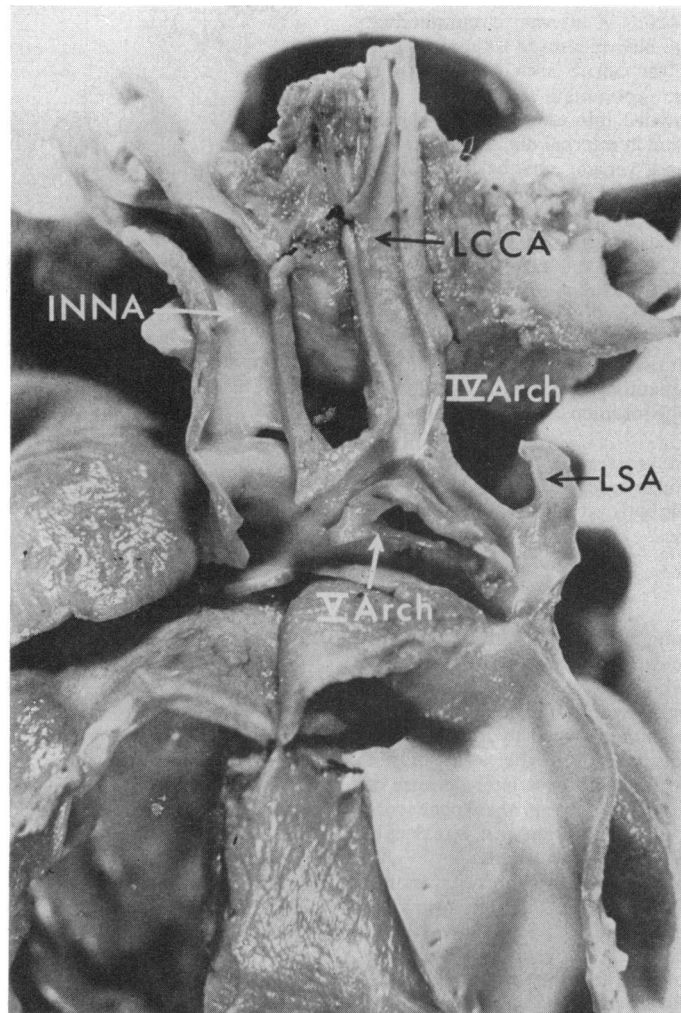


FIG. 5 Case 2. Specimen of the area of the aortic arch showing the two arches clearly separated (IV and V). The innominate artery (INNA), left common carotid artery (LCCA), and left subclavian artery (LSA) arise from the left fourth aortic arch. The localized coarctation of the aorta immediately distal to the left subclavian artery and the persistent ductus arteriosus is noted.

(1913) and in the chick embryo, Buell (1922) describe a left fifth aortic arch forming a vascular channel caudal to the fourth aortic arch. Congdon (1922) demonstrated the presence of endothelial sprouts arising from the aortic sac and the descending aorta which were thought to be the ends of the fifth aortic arches in the human embryo. Finally, Van Praagh and Van Praagh (1969) presented the first case of a persisting left fifth aortic arch in a human postmortem heart.

The two cases presented in this report add further support to the developmental concept that a fifth arch can occur in man. Case 2 is particularly important because the two arches were clearly separated by a gap and each arch was surrounded by an adventitial sheath.

Case 1 is, to the best of our knowledge, the first persistent left fifth aortic arch to be recognized during life. As suggested by Van Praagh and Van Praagh (1969), the diagnosis was made from the selective angiocardigram of the aortic arch system obtained at cardiac catheterization (C. A. F. Moes and T. Izukawa, 1973, unpublished observations). Awareness that the entity exists is important, as alternative explanations for the odd angiographic appearance can be produced. One possible explanation is that the misplaced vessel is an aberrant brachiocephalic artery, usually a right subclavian artery originating abnormally from the descending aorta. A routine chest x-ray and haemodynamic data from cardiac catheterization will not detect the anomaly. A normal oesophagogram will rule out both the existence of an aberrant right subclavian artery and a double aortic arch (fourth).

In all three cases reported to date, the persisting fifth aortic arch was on the left side and originated from the aortic lumen at or distal to the origin of

the innominate artery and rejoined the aortic lumen at the origin of the left subclavian artery inferiorly. All three cases had other abnormalities of the heart or great vessels. The occurrence of an isolated persisting left fifth aortic arch has not yet been reported.

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